

Ultomiris™ (ravulizumab-cwvz)

When requesting Ultomiris (ravulizumab-cwvz), the individual requiring treatment must be diagnosed with an FDA-approved indication and meet the specific coverage guidelines and applicable safety criteria for the covered indication.

FDA-approved Indications

Ultomiris is indicated for:

- The treatment of adult patients with paroxysmal nocturnal hemoglobinuria (PNH).
- The treatment of patients with atypical hemolytic uremic syndrome (aHUS) to inhibit complement-mediated thrombotic microangiopathy.

Coverage Guidelines

Paroxysmal nocturnal hemoglobinuria (PNH)

The individual must meet **all** of the following criteria for initial authorization:

- The diagnosis of PNH has been confirmed by peripheral blood flow cytometry results showing the absence or deficiency of glycosylphosphatidylinositol (GPI)-anchored proteins on at least 2 cell lineages;
- The individual is 18 years of age or older; AND
- Ultomiris is prescribed by or in consultation with a hematologist.

For re-authorization:

- An individual is continuing to receive benefits from Ultomiris (e.g., stabilization of hemoglobin levels, decreased transfusion requirement or transfusion independence, reduction in hemolysis).

Approval duration (initial): 6 months

Approval duration (renewal): 12 months

Atypical Hemolytic Uremic Syndrome (aHUS)

The individual must meet **all** of the following criteria for authorization:

- The individual does not have Shiga toxin *Escherichia coli* related hemolytic uremic syndrome; AND
- Ultomiris is prescribed by or in consultation with a nephrologist.

Approval duration: 12 months

Dosing Recommendations

Ultomiris is only administered as an intravenous infusion.

Paroxysmal nocturnal hemoglobinuria (PNH)

Administer loading dose once, followed 2 weeks later by maintenance dose. Administer maintenance dose once every 8 weeks.

Body Weight Range (kg)	Loading Dose (mg)	Maintenance Dose (mg)	Maintenance Dosing Interval
Greater than or equal to 40 to less than 60	2,400	3,000	Every 8 weeks
Greater than or equal to 60 to less than 100	2,700	3,300	Every 8 weeks
Greater than or equal to 100	3,000	3,600	Every 8 weeks

Atypical hemolytic uremic syndrome (aHUS)

Administer loading dose once, followed 2 weeks later by maintenance dose. Administer maintenance dose once every 8 weeks or every 4 weeks depending on body weight.

Body Weight Range (kg)	Loading Dose (mg)	Maintenance Dose (mg)	Maintenance Dosing Interval
Greater than or equal to 5 to less than 10	600	300	Every 4 weeks
Greater than or equal to 10 to less than 20	600	600	Every 4 weeks
Greater than or equal to 20 to less than 30	900	2,100	Every 8 weeks
Greater than or equal to 30 to less than 40	1,200	2,700	Every 8 weeks
Greater than or equal to 40 to less than 60	2,400	3,000	Every 8 weeks
Greater than or equal to 60 to less than 100	2,700	3,300	Every 8 weeks
Greater than or equal to 100	3,000	3,600	Every 8 weeks

References

1. Ultomiris™ injection [prescribing information]. New Haven, CT: Alexion Pharmaceuticals, Inc.; October 2019.
2. Brodsky RA. Paroxysmal nocturnal hemoglobinuria. *Blood*. 2014;124(18):2804–2811.
3. Soliris® injection [prescribing information]. New Haven, CT: Alexion Pharmaceuticals, Inc.; November 2019.
4. Campistol JM, Arias M, Ariceta G, et al. An update for atypical haemolytic uraemic syndrome: diagnosis and treatment. A consensus document. *Nefrologia*. 2015;35:421–447.
5. Genetics Home Reference. Atypical hemolytic-uremic syndrome. National Institutes of Health (NIH). Available at: <https://ghr.nlm.nih.gov/condition/atypical-hemolytic-uremic-syndrome#sourcesforpage>. Accessed on October 31, 2019.
6. Ravulizumab-cwvz (Ultomiris®). In: DRUGDEX® System Micromedex. Greenwood Village, CO: Thompson Reuters (Healthcare) Inc. [Updated; November 12, 2019; Accessed December 23, 2019].